# Two clinicopathological cases of a dominantly inherited, adult onset orthochromatic leucodystrophy



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| Titre | Two clinicopathological cases of a dominantly inherited, adult onset orthochromatic leucodystrophy |
| Type de publication | Article de revue |
| Auteur | Letournel, Franck [1], Etcharry-Bouyx, Frédérique [2], Verny, Christophe [3], Barthelaix, Annick [4], Dubas, Frédéric [5] |
| Editeur | BMJ Publishing Group |
| Type | Article scientifique dans une revue à comité de lecture |
| Année | 2003 |
| Langue | Anglais |
| Date | Mai 2003 |
| Numéro | 5 |
| Pagination | 671-3 |
| Volume | 74 |
| Titre de la revue | Journal of neurology, neurosurgery and psychiatry |
| ISSN | 0022-3050 |
| Mots-clés | Adult [6], Age of Onset [7], Brain [8], Female [9], Genes, Dominant [10], Humans [11], Leukodystrophy, Metachromatic [12], Magnetic Resonance Imaging [13], Male [14], Middle Aged [15], Pedigree [16], Phenotype [17] |
| Résumé en anglais | Leucodystrophies of orthochromatic type are a heterogeneous group that occur mainly in childhood and have no known enzyme deficiency. We report here the clinicopathological features of a new family of orthochromatic leucodystrophy with three main characteristics: a probably autosomal dominant inheritance; two phenotypes based on age of onset; and very few abnormalities of white matter on MRI findings in one case. The first patient, aged 58 years, had frontal dementia and epilepsy; the second, aged 38 years, had motor signs and dementia, but no epilepsy. The histopathological features of our two cases were leucodystrophy of orthochromatic subtype. However, the radiological features (MRI and mostly FLAIR sequences) of the first case did not suggest leucodystrophy. |
| URL de la notice | http://okina.univ-angers.fr/publications/ua15662 [18] |
| DOI | 10.1136/jnnp.74.5.671 [19] |
| Lien vers le document | http://jnnp.bmj.com/content/74/5/671.full [20] |
| Titre abrégé | J. neurol., neurosurg. psychiatry |
| Identifiant (ID) PubMed | 12700318 [21] |
| PubMed Central ID | PMC1738413 |

**Liens**
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[2] http://okina.univ-angers.fr/frederique.bouyx/publications
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